

HEMIVÉRTEBRA: A CASE REPORT

Hemivertebrã: Relato de caso

Carla Moreira da Costa¹, Isabela Bianca Rodrigues Fernandes¹, Eriston Vieira Gomes^{2*}

RESUMO

A coluna vertebral forma o centro do esqueleto do corpo humano, é composta por 33 vértebras divididas em região cervical, torácica, lombar, sacral e coccígea. A hemivértebra (HV) é uma má formação dessa estrutura, sendo a escoliose congênita a complicação mais recorrente. Essa condição é caracterizada como uma curvatura anormal lateral da coluna. O prognóstico da HV é relacionado a região de acometimento e complicações associadas. A etiologia é subsequente a um erro no centro de condrificação da vértebra no período gestacional por deslocamentos incoerentes de células na embriogênese. O Tratamento tardio pode trazer deformidades da coluna e gerar respostas anatômicas e sistêmicas alteradas.

Palavras-chave: Hemivértebra; Escoliose; Escoliose Congênita; Coluna Vertebral

ABSTRACT

The spine forms the center of the human skeleton, and is composed of 33 vertebrae divided into cervical, thoracic, lumbar, sacral, and coccygeal regions. Hemivertebrã (HV) is a malformation of this structure, with congenital scoliosis being the most recurrent complication. Scoliosis is characterized by abnormal lateral curvature of the spine. The prognosis of HV is related to the region of involvement and the associated complications. The etiology is subsequent to an error in the center of chondrification of the vertebra during gestation by incoherent displacement of cells during embryogenesis. Late treatment can lead to spinal deformities and altered anatomical and systemic response. This study aimed to describe a case of hemivertebrã and its evolution.

Keywords: Hemivertebrã; Scoliosis; Congenital Scoliosis; Spine;

1. Graduanda em Medicina, Faculdade Morgana Potrich (FAMP). Mineiros- GO, Brasil.

2. Doutorado em Bioquímica na Faculdade de Medicina de Ribeirão Preto da Universidade de São Paulo (FMRP-USP). Docente na Faculdade Morgana Potrich (FAMP), Mineiros-Go, Brasil.

*Autor para Correspondência. E-mail: eristonvieira@fampfaculdade.com.br



INTRODUÇÃO

The vertebral column forms the center of the human skeleton and is formed of bones called vertebrae (Mahadevan, 2018). Its functions are to support the body, protect the nervous system, and perform movements. It is composed of 33 vertebrae, divided into seven cervical, 12 thoracic, five lumbar, five sacral, and four coccygeal vertebrae (DeSai et al., 2021). The hemivertebra (HV) is a formation deformity on one side of the vertebral body and is considered one of the main congenital anomalies of this structure (Wen et al., 2018). The spine can be affected by HV throughout its length; however, this is less frequent in the cervical region (Dayer et al., 2017) and more common in the thoracic and lumbar regions (Johal et al., 2016). The occurrence of VH ranges from one to ten in every ten thousand live births, and some studies indicate a higher frequency in males; however, other studies have shown that the occurrence is equal between the two sexes (Johal et al., 2016). The incidence among families varies between 1% and 5% (Bao et al., 2020).

The etiopathogenesis of HV stems from an error in the vertebral chondrification center (Yulia et al., 2020). In the fourth week of gestation, somite cells are displaced and the mesenchymal spine is formed, which becomes important for vertebral chondrification centers (Blevins et al., 2018). Somite formation features intertwined embryonic sequences formed by a single portion of the non-segmented paraxial or presomitic mesoderm (McColl et al., 2018). With the formation and maturation of somites, differentiation begins. Among these differentiations is sclerotome (Maroto et al., 2012). The incoherent displacement with irregular directions and movements of somite cells corroborate the unusual development of HV (Dayer et al., 2017). Environmental causes, such as carbon monoxide inhalation, gestational diabetes, and the use of antiepileptic drugs, may favor this anomalous event (Bao et al., 2020).

HV presents different consequences according to the growth of the spine, thus depending on the type, growth capacity of the individual, and place of involvement. HV has three main complications: scoliosis, hyperlordosis, and hyperkyphosis, with scoliosis being the most common (Bao et al., 2020). Scoliosis is characterized by abnormal lateral curvature of the spine (Depaola and Cuddihy, 2020). The congenital subtype of scoliosis has HV as the main etiology (Depaola and Cuddihy, 2020; Yang et al., 2020). An increase in the anterior curve of the spine is characterized by hyperkyphosis, while lordosis is defined by an increase in the posterior curvature, both analyzed in the sagittal plane (Lam and Murkhdomi, 2021).

As a result of the deformity arising from the HV spine

growth is unbalanced. In turn, the adjacent nonpathological vertebrae are overloaded and begin to develop asymmetrically. Thus, congenital abnormalities of the spine can lead to adverse events in cardiorespiratory function, which is the main morbidity associated with this alteration, functional deformity in the musculoskeletal area, and psychological suffering in the patient. In turn, HV resection is the gold standard for congenital scoliosis caused by HV (Ouellet, 2011; Bao et al., 2021; Weiss et al., 2021). Prognosis and post-surgical complications include implants displaced out of the inserted site, rod breakage, site infection, healing difficulties, and peripheral nerve and spinal cord injury, causing neurological dysfunction (Zhang and Zhang, 2020).

RESULTS AND DISCUSSION

A 12-year-old female student was admitted to Hospital Santa Luzia in the city of Luziânia, Goiás, in 2008 for the first medical consultation with complaints of progressive enlargement of the posterior thoracic region. This increase was more relevant when her body was bent forward, and she reported feeling pain and discomfort when standing or sitting for long periods. In the patient's report of her neuropsychomotor development, no paresthesia, plegia, or difficulty in walking were found, and only falls from her own height with no apparent cause were noted. The patient denied the occurrence of any other diseases. At the time of the consultation, physical examinations were performed, such as the Adams test, the distraction test, and the patellar and Babinski reflexes, and an anteroposterior and profile X-ray were requested (Fig. 1). Consequently, the patient was advised to seek an orthopedic spine specialist. At this second evaluation, the specialist requested magnetic resonance imaging (MRI) of the spine (Fig. 2), and the diagnosis of non-segmented hemivertebra at T12 was made.



Figure 1. Radiograph in profile. Evidence of enlargement in the posterior thoracic region (a) is visible.

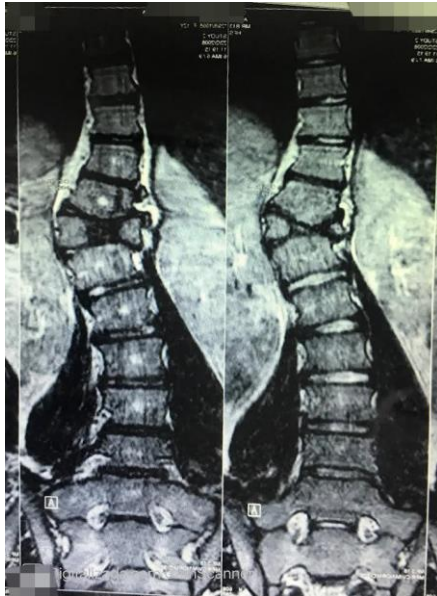


Figure 2. Nuclear magnetic resonance image evidencing the VH at T12 in the anterior position (b).

The definitive surgical technique for treating HV is hemivertebrectomy (Barik et al., 2021), which can be performed using an anterior, posterior, or combined approach. The posterior approach to hemivertebral excision is the most commonly used method, with bilateral screw fixation in the pedicle region of the short segment of the vertebrae. Spinal nerve monitoring is performed during the operation, and the patient is placed in the prone position under general anesthesia (Wang et al., 2021). Therefore, surgical intervention was used, considering that noninvasive clinical measures do not have satisfactory long-term effects.

Therefore, the patient and her parents were informed of the therapeutic option, and the procedure was divided into two surgeries 10 days apart. In the first surgery, the strategy consists of removing the hemivertebra and performing a bone graft at the site (important as a "bridge" to connect the operated vertebrae). In the second surgery, spinal arthrodesis is performed to stabilize to the spine by immobilizing some of its segments.

Separating the surgery into two phases provides a more favorable prognosis without major complications. In contrast, performing the entire procedure at once could provide the benefit of shorter hospital stay and faster recovery, but with longer surgical procedure time, thus increasing the risk of interurrences, such as spinal nerve lesions, and consequently, damage to neuro-motor processes. After presenting therapeutic options to the parents, the procedure was performed in two phases. Both surgeries were performed at the Hospital de Acidentados Clínica Santa Isabel in the city of Goiânia, GO.

On March 2008, the first surgical procedure was performed, using two left lateral incisions to remove the hemivertebra and a bone graft at the site using part of the left iliac bone (Fig. 3). The surgical process lasted six hours, with an interurrence of hemothorax; therefore, a water-sealed drain was used. The problem was resolved after 4 days. The patient experienced no complaints in her general state until day 10 after the first surgery and remained in dorsal decubitus for the entire period, performing all of her activities and needs while in her own bed. Medications were prescribed, morphine every eight hours for the first three days and tramadol every eight hours for subsequent days in case of pain.

A second surgery was performed nine days later. A posterior-approach spinal arthrodesis was performed, in which two titanium rods were inserted and 16 screws were placed from T1 to L4 (Figs. 4 and 5). The procedure lasted eight hours, and it was necessary to perform the awakening test for a motor evaluation, which indicated no changes. There were no interurrences during this second procedure, and the patient was hemodynamically stable. On the second postoperative day, she took her first steps without motor or sensory alterations. The same medications were prescribed as for the first surgery, and the surgical incision did not involve seroma, broken stitches, or infection.

On April 2, 2008, the patient was discharged from the hospital and returned after 15 days for postoperative evaluation and stitch removal. Fifteen days later, she was in a good general state with no pain and a satisfactory recovery.



Figure 3. Anteroposterior hip radiograph taken after removal of the bone graft (c).



Figure 4. Anteroposterior radiograph taken after posterior spinal arthrodesis (d).



Figure 5. Lateral radiograph taken after posterior spinal arthrodesis (e).

CONCLUSION

Early diagnosis favors a better prognosis, and is performed during pregnancy, by means of ultrasonography.

However, diagnosis is difficult because it is an operator-dependent test and the mother does not follow up with regular prenatal care. The pathophysiological aspects arising from this lack of early diagnosis leads to a worsening of the abnormal curvature of the patient's spine. It is the precocity of the diagnosis that will give us the best results in the treatment. Not only in the anatomical part, but also in the psychological part of the patient and his parents. From the very beginning of their diagnosis, they will be able to better conduct all the procedures that they may face, surgical and non-surgical. The embryological and etiological aspects are the starting factors for this HV pathology. Posterior hemivertebra resection is a safe and efficient intervention in cases of congenital hemivertebral scoliosis. Early intervention allows development without local deformities and without secondary curves, contributing to normal spinal growth. Therefore, the present report was limited to the description of a clinical case about congenital scoliosis due to a HV. It provided relevant data on the clinical features, possible causes, complications, early diagnosis, prognosis, and treatment. Moreover, as in any other pathology that is susceptible to mutations, whether embryological or even anatomical, we emphasize the continued need for further research on the disease mentioned throughout the article.

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